PEDIATRIC NEWS

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Housestaff Puzzler

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SS is a 4 year-old female who was brought to the emergency room for "blood in her underwear". Her parents reported that she had been at a friend's birthday party and had consumed a large amount of food and liquid. She began complaining of severe left-sided abdominal pain, and was noted to have blood in her underwear after using the restroom. They were unsure of whether the blood was from her vagina or her rectum, but felt it was probably the former as she denied having a bowel movement.

60% of all neonatal hydronephrosis will be due to UPJ obstruction.

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In this edition

The DoD/VA Asthma Clinical Practice Guideline A Sound Basis for Asthma Management

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Ok, have you heard of the DoD disease management guidelines? Asthma and back pain were the first of these MEDCOM sponsored guidelines to be completed and implemented, and there are several more on the way. There are many ways to learn how to treat diseases, and currently one popular method is to use evidence-based clinical practice guidelines. Whether guidelines can influence and improve care is still subject to debate, but I think we can all agree that a well-constructed guideline can serve as a sound reference for teaching good medicine. The DoD Asthma guideline is an excellent reference, and my purpose in this article is to introduce you to that guideline and its key points.

Background and history of asthma guidelines

It became obvious in the late 1980's that as a profession we were not doing so well managing asthma. Asthma has always been amongst the most common chronic illnesses of children, and by 1990 there was a significant increase in prevalence, severity, and mortality.[1,2] In addition, asthma management varied widely at all care levels. This prompted the NIH to form a consensus panel, which then wrote the first version of the National Heart Lung & Blood Institute Guidelines for the Diagnosis and Management of Asthma (NHLBI Asthma Guidelines). This excellent and informative document was published in 1991 as a monograph and mailed to practically every physician in the country. It sat on office shelves, found its way

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Update in Childhood Epilepsy

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Seizures and Epilepsy are common neurologic issues which the general pediatrician faces both in the outpatient clinic as well as the emergency room. Over the past decade numerous changes regarding the diagnostic evaluation, therapeutic intervention, and the medications available for treatment have developed. This article will attempt to summarize currently accepted treatment protocols for the new millennium.

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Puzzler

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Her parents denied any preceding URI symptoms, fever, weight loss, and prior hematochezia, melena, or hematuria. She had a history of recurrent abdominal pain for which she presented to the pediatrician and ER multiple times, often associated with special occasions when she ate and drank

"too much". These episodes were felt to be indigestion and they resolved spontaneously, so no further workup was done.

Her past medical history is significant only for the recurrent abdominal pain. She had no surgeries and was not taking any medications. Her family history was non-contributory.

Physical Exam

Vitals HR 92, BP 95/56, RR 22, SaO2 99% RA

Gen: thin AA female in no apparent distress, sitting comfortably in her father's arms

HEENT: WNL

CV: WNL, hemodynamically stable

Resp: WNL

GI: soft, diffusely tender abdomen with 2x3 cm firm mass palpable in the left upper quadrant, normal bowel sounds, no HSM

GU: normal female, no evidence of urethral irritation or bleeding

Rectal: normal tone, small amount of soft stool in vault, guaiac negative

Skin: WNL, no bruising or petechiae

*The most common oncologic cause of hematuria and an abdominal mass is **Wilms' tumor**.*

Laboratory Studies

CBC: WBC 10.6, Hgb 12.3, Hct 34.6, plt 326

Chem: Na 139, K 3.8, Cl 104, HCO3 24, BUN 16, creat 0.3, glucose 73

Coags: PT 12, PTT 22.1

UA (cath): yellow, SG 1.025, pH 6.5, 3+ ketones, neg glucose/protein, neg nitrite, neg LE, 3+ blood, 20-29 RBC, 15-19 WBC, 1-4 epi, trace bacteria, no casts

Urine culture: pending

Course

SS was admitted to the pediatric ward for further evaluation of her left-sided abdominal mass and hematuria. A renal and bladder ultrasound was performed. The study was significant for marked left sided hydronephrosis (left kidney 9.1 cm, right kidney 7.3 cm) and a dilated left renal pelvis which ended abruptly. The diagnosis of left ureteropelvic junction (UPJ) obstruction was made. Pediatric urology was consulted. They further imaged her with a VCUG (negative) and diuretic renography (T1/2 pre-lasix of 37 minutes, which reduced to 4 minutes post-lasix). Because her renal function was minimally damaged and she was symptomatic from her UPJ obstruction, she was taken to the OR three months later for elective open pyeloplasty. She is doing well post-operatively with stable renal function and no further symptoms.

Discussion

This case is interesting from two standpoints:1) discussion of the differential diagnosis of gross hematuria in a child, and 2) further evaluation of UPJ obstruction itself as a cause of gross hematuria.

Gross hematuria

Gross hematuria is defined as visible red urine which is determined to be caused by RBCs seen on microscopic analysis. Most clinicians further delineate gross hematuria into glomerular or non-glomerular by the presence or absence of protein. A discussion of microscopic, glomerular, and false-positive hematuria is outside the scope of this discussion, which will concentrate on gross hematuria only. The differential diagnosis of pediatric gross hematuria includes:

1) Anatomical abnormalities: meatal stenosis with ulcer, renal vein/arterial thrombosis, posterior urethral valves (PUV), polycystic kidney disease, vesicoureteral reflux (VUR), UPJ/UVJ obstruction, renal arteriovenous fistulae
2) Cancer: Wilms' tumor, mesoblastic nephroma, rhabdomyosarcoma
3) Trauma: child abuse, traumatic catheterization, blunt abdominal trauma, foreign body
4) Stones: nephrolithiasis, hypercalciuria
5) Factitious: menstruation, hematochezia, exercise-induced perineal irritation
6) Infectious: UTI
7) Hematologic: sickle cell disease/trait, clotting disorders

This patient's evaluation for gross hematuria was guided by the presence of an abdominal mass, which led to radiographic workup. Obviously, the evaluation of a patient with hematuria should be tailored to their clinical presentation.

UPJ Obstruction

UPJ obstruction specifically as a cause of gross hematuria is rare now that the diagnosis is usually made antenatally. Out of all cases of antenatal hydronephrosis (up to 2.0% of all prenatal ultrasounds),

about 60% of those neonates who have true hydronephrosis on a post-natal ultrasound will have UPJ obstruction. The remaining 40% is divided among other congenital lesions like VUR and PUV.

UPJ obstruction is more common in males at approximately 3-4:1 incidence. Left-sided lesions predominate at about 65% of all cases. UPJ obstruction is bilateral in up to 40% of cases, with the majority of those being asymmetric (one side worse than the other). Another well-known association with UPJ obstruction is that of contralateral multicystic dysplastic kidney (MCDK), which likely represents bilateral UPJ obstruction with the MCDK being the most severely affected kidney.

In one prospective study, the causes of UPJ obstruction were intrinsic stenosis (75%), anomalous insertion of the ureter (7%), fibrous bands causing external compression (3%), and anomalous blood vessels crossing over the ureter and causing external compression (11%). The theory behind the most common cause (intrinsic stenosis) is that of a *hypoplastic adynamic ureteral segment* which carries urine at low workloads but cannot adjust to high-volume periods. Our patient suffered from this cause, which led to the interesting association of her abdominal pain with birthday parties and other occasions in which she drank large volumes of liquid and exceeded the limit of her left ureter's capacity.

*A common cause of factitious hematuria in an older female child is **menstruation**.*

Clinical presentation of UPJ obstruction outside of the neonatal period commonly includes UTI's, abdominal pain, abdominal masses, hematuria, and GI symptoms. Less common presentations include failure to thrive, anemia, and hypertension. The VATER syndrome is associated with UPJ obstruction, and a renal ultrasound should be obtained in patients with any of those malformations.

Diagnosis in the neonatal period should be treated like any other antenatal hydronephrosis with a post-natal ultrasound and a VCUG. Similarly, older children with concerning symptoms should receive the same evaluation. Together these tests should diagnosis those patients with true UPJ obstruction. These patients should then be referred to pediatric urology for surgical management. The surgical options include balloon dilation, and open pyeloplasty (essentially reconstruction and reattachment of the ureter after removal of the narrow segment) or endopyelotomy (balloon dilation and cutting with stent placement). The goal in early surgical management is to prevent renal parenchymal damage and symptoms such as our patient experienced.

In summary, gross hematuria with an abdominal mass is unlikely to be UPJ obstruction in the older child given the frequency of antenatal ultrasounds and prenatal diagnosis. However, it should be on the differential of any child with hematuria or recurrent abdominal pain. Consultation with the pediatric urologist is essential after the diagnosis is made for proper surgical management with the goal of preserving renal function.

The majority of all UPJ obstruction is located on the left side.

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DOD/VA Asthma

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to the trashcan, and was occasionally read. However, it impacted little on asthma care. In 1997, a second version was published, accompanied by a more rigorous implementation strategy.[3] This did impact on asthma care but not extensively.[4] Shortly thereafter, the Veteran's Administration adapted the '97 NHLBI guidelines, and put them into an algorithm/flow chart format. In 1998, LTG Blank, the Army Surgeon General, instituted a program to standardize care across the MEDCOM, and part of this program was to produce and implement clinical practice guidelines. This prompted the first Army guideline meeting in March 1998. During the next year, a large group of primary care providers and

subspecialists met to adapt the VA asthma guidelines for the military. This culminated in the formation of algorithm-based practice guidelines for patients of all ages with asthma. All three military services, the VA, and the Public Health Service have adopted this guideline. A rigorous implementation process began in August 2000 and continues to this day. Thus, each of you should have heard about the DoD asthma guideline, though I suspect few have actually read it through.

The DoD/VA clinical practice guideline for asthma:

The asthma and the other DoD/VA guidelines can be accessed at the MEDCOM Quality Assurance web page: www.cs.amedd.army.mil/qmo. There is a PDF version that can be printed out and also a web navigable one. There are links to provider and patient educational tools, reminder cards (key points) and other asthma information. The guideline guide is separated into two parts, one guideline for children < 6-years-old who cannot perform spirometry and another for older children and adults who can perform spirometry. Each of these two guidelines has four components – Initial Diagnosis and Management, Long-term Management, Acute/Emergency Management, and Telephone Management. Each component has an algorithm/flow diagram accompanied by text annotations that highlight key information and provide the level and grade of evidence. While, as with most large guidelines, you may find it cumbersome to view and use this asthma guideline, it really contains all the information required for you to provide appropriate care to your patients with asthma. While there is considerable detail and information in the asthma guideline, much of it can be distilled down to important "key" points. In the remaining portion of this article I will highlight the key points of the first 2 components of the guidelines, Initial Diagnosis and Management and Long-Term Management, as well as discuss some of the differences between the younger child and older child/adult guidelines. The telephone management is self-explanatory, and there is not enough space to cover the emergency management of acute asthma.

Make the diagnosis of asthma. It's common.

Initial diagnosis and management – key points

1. Consider asthma in the differential diagnosis of any child that presents with persistent or recurrent respiratory symptoms. The guideline provides a differential diagnosis as well as common presenting signs and symptoms. If you think about asthma and know how it presents, then you will be much better able and, hence, more likely to diagnose it.

2. Use spirometry to help make the diagnosis. The diagnosis algorithm for the older children and adults is "spirometry driven." Once asthma is suspected then the patient should perform spirometry (PFTs). Many children with asthma will have normal spirometry, but if their values are abnormal, then this really helps in making the diagnosis. Peak flow measurements cannot replace spirometry in the diagnosis process. Thus, all providers caring for children must become familiar with how to obtain spirometry results and know how to interpret them.

3. Use trials of asthma medications and patients' responses to them as an aid to diagnosis. If spirometry is normal but you suspect asthma based on history and physical findings then consider a trial of asthma medications. Usually this is a trial of a bronchodilator, e.g. albuterol, but it could include a short course of PO corticosteroids or several weeks of a controller medication. The DoD asthma guideline provides you with guidance on how to prescribe trials of medication.

Classify asthma severity.

Long-term asthma management

1. Classify asthma severity. This is all-important. If you do not classify the severity of the patient's asthma then it will be difficult for you to devise the appropriate management plan. Use frequency/severity of signs and symptoms, medications required to control symptoms, and objective measures of airways obstruction (peak flow, spirometry) to categorize your patients as mild intermittent, mild persistent, moderate persistent, or severe persistent. Patients often bounce back and forth across these categories, so re-categorize at each follow-up visit.

2. Treat patients based upon their asthma severity classification. Patients with persistent asthma require controller medications. The guideline lists the medications and their dosages for each of the severity categories. Inhaled corticosteroids are still the first line drug for most children with persistent asthma, though cromolyn, leukotriene receptor antagonists, and long-acting beta$_2$ agonists all have their roles.

3. Educate patients about their asthma. While education is always a good thing, the strategies that have the best backing from the literature are a written action plan and an accessible point of contact. Action

Prescribe controller medications for patients with persistent asthma.

plans can be quite detailed, but even simple ones are beneficial. A written plan that lists the patient's medications, tells what to do if increased symptoms arise, and notes the provider's phone number are minimum requirements.

4. Assess for triggers, and counsel on trigger avoidance as you deem necessary. The guideline lists the common triggers of asthma and discusses in some detail allergy evaluation and allergen control measures. You should evaluate most children ³ 5-years-old who have moderate or severe persistent asthma for common aeroallergens using either RAST or skin prick testing. All children younger than 5-years-old, even those with moderate or severe persistent asthma, and older patients with mild persistent asthma should be considered for allergy testing on a case by case basis.

5. All patients with asthma need to have a primary care manager (PCM) and regular follow-up. Update actions plans, re-educate, and review inhaler instruction at each visit.

Differences between the guideline for younger patients who cannot perform spirometry and the one for older patients who can.

1. The guideline for young children and the one for older children/adults are remarkably similar.
2. The main difference is that younger children cannot perform spirometry, so diagnosis and asthma severity are primarily based on history, signs, and symptoms.
3. Allergies do not play as major a role in children < 5-years-old and are not commonly a problem in children < 2-years-old.

There have been many articles published on asthma guideline use, implementation, and effectiveness. Most studies show that there are many barriers to guideline use, that many providers still deviate from the standards set by the NHLBI and DoD asthma guidelines, and that many pediatricians find guidelines too complicated.[5-7] Yet, if a guideline is done well and you know what it contains, then you are a long way towards practicing good medicine. That's what guidelines are all about. You use them to implement best practices. If you take the time to learn what is emphasized in the DoD/VA asthma guideline and practice what it preaches, then you will successfully manage the majority of your patients with asthma.

Order spirometry for children who can perform this test.

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Epilepsy

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To review some pertinent statistics illustrating the impact of seizures in pediatrics:

*Seizures occur in 3-5% of the pediatric population

*Febrile seizures occur in 3% of the pediatric population

*Epilepsy occurs in .5-1% of the population with 60% of cases beginning in childhood (translating into 30,000 new pediatric cases annually)

First Simple Febrile Seizure

The *American Academy of Pediatrics* published a practice guideline statement in May 1996 making recommendations regarding the diagnostic evaluation of children presenting with a first simple febrile seizure. The four diagnostic procedures (blood studies, lumbar puncture, neuroimaging, and electroencephalogram) routinely considered and performed in various combinations were reviewed with the following recommendations made.

Blood Studies

-recommend glucose level in a child with prolonged obtundation,

not as part of the routine evaluation

-recommend CBC as part of the evaluation of fever, not as part of the routine evaluation

-electrolytes, calcium, phosphorous, and magnesium not recommended as part of the routine evaluation

Lumbar puncture

-recommended in infants younger than 12 months

-recommended in children older than 18 months with meningeal signs and symptoms

-recommended in children having received prior antibiotic therapy

-consider performing LP in children between ages 12 and 18 months based on history/exam

Neuroimaging

-not recommended as part of the routine evaluation

Electroencephalogram

-not recommended as part of the routine evaluation

Regarding therapy, phenobarbital is no longer recommended as prophylactic treatment against febrile seizures due to the recognized cognitive impairment associated with long term exposure to the drug. Therefore, in cases of patients with recurrent febrile seizures or a history of febrile seizures progressing to febrile status epilepticus, rectal valium may be prescribed for acute abortive therapy. The dose of valium for rectal administration is .3mg/kg to be given at the onset of seizure activity. *DIASTAT* is diazepam in gel form prepared in 2.5mg and 5.0mg syringes for single use dosing.

First Afebrile Seizure

The *American Academy of Neurology* published a practice guideline in September 2000 making recommendations on the diagnostic evaluation of a child presenting with a first afebrile seizure. The same four diagnostic procedures discussed above were also reviewed with the following recommendations made.

Blood Studies

-not recommended for routine evaluation in children who are greater than 6 months and whose seizure spontaneously resolved with return of baseline mental status at time of evaluation

-individual clinical circumstances (ie. failure to return to baseline mental status, emesis, etc) may direct the physician to order specific laboratory testing

-toxicology screening should be considered if drug exposure or substance abuse is suspected

Lumbar Puncture

-recommended only if a concern for meningitis or encephalitis exists

-imaging of the head recommended prior to LP if increased intracranial pressure suspected

Neuroimaging

-emergent imaging (non-constrast CT of the brain) recommended in a child who exhibits postictal focal neurologic deficit (Todd's paralysis), or failure to return to baseline mental status within several hours after the seizure's resolution

-non-emergent imaging (magnetic resonance of the brain) recommended in a child with cognitive or motor impairment, abnormal finding on neurologic examination, children under age 1 year, a seizure of partial (focal) onset, and an EEG abnormality which does not represent a benign partial epilepsy of childhood or primary generalized epilepsy.

Electroencephalogram

-recommended for all patients after experiencing first afebrile seizure

-a sleep-deprived study employing hyperventilation and intermittent photic stimulation performed within 24-48 hours of the seizure will increase yield of identifying abnormalities.

It is accepted practice that anti-epileptic drugs are not initiated for seizure prophylaxis after a first afebrile seizure given that the majority of patients (~70%) will not develop epilepsy.

Discontinuing Anti-Epileptic Medications in Patients with Epilepsy

The *American Academy of Neurology* published a practice guideline in September 1994 establishing diagnostic criteria for discontinuing anti-epileptic medication in patients with epilepsy who had been seizure free for an extended period. If a child fits the profile listed below, a 69% chance for successful withdrawal is expected. Thus, a relapse rate of 31% should be included in counseling families.

*Seizure-free 2-5 years on anti-epileptic drug therapy

*Epilepsy manifested by a single type of partial or generalized seizure

*Normal neurologic examination and normal IQ

*EEG normalized with treatment

New Generation Anti-Epileptic Drugs

1993 marked the release of new anti-epileptic medications after a 15 year hiatus. Since then, nine drugs have entered the market primarily

indicated for adjunctive use in patients whose epilepsy is not controlled with a single drug. It is important to note that these new AEDs are highly effective and, in this author's opinion, will eventually replace older,established medications (carbamazepine, valproic acid, phenytoin, phenobarbital). The reason for this is the broader spectrum of action of the new generation AEDs. The previous generation drugs acted to control neuronal hyperexcitability by reducing levels of the excitatory neurotransmitter glutamate by blocking pre-synaptic Na+ channels (CBZ, VPA, PHT) or increasing the levels of the inhibitory neurotransmitter GABA by stimulating the post-synaptic GABA receptors. Today's newer AEDs act to control epilepsy by decreasing excitatory neurotransmission and increasing membrane hyperpolarization through their action on calcium channels, NMDA receptors, and AMPA/kainate receptors, as well as having impact on Na+ channels and GABA receptors.

The following is a list of the new AEDs which the general pediatrician may encounter.

Of note, unlike the previous generation AEDs, serum drug levels of the new medications are not routinely obtained. Therefore, in alphabetical order:

* Felbamate (*Felbatol*) 45-60mg/kg/day divided three times daily

* Gabapentin (*Neurontin*) 30-100mg/kg/day divided three times daily

* Lamotrigine (*Lamictal*) 1.0-15mg/kg/day divided twice daily

* Levetiracetam (*Keppra*) 40-60mg/kg/day divided twice daily

* Oxcarbazepine (*Trileptal*) 20-45mg/kg/day divided twice daily

* Tiagabine (*Gabatril*) .6-1.0mg/kg/day divided three times daily

* Topiramate (*Topamax*) 5-9 mg/kg/day divided twice daily

* Zonisamide (*Zonegran*) 4-8 mg/kg/day once daily or divided twice daily

Changes in the Treatment of Status Epilepticus

The two significant changes in the treatment of status epilepticus over the past 5 years has been the development of the drug fosphenytoin (*Cerebryx*) and the market shortage of phenobarbital. Fosphenytoin is a water-soluble phosphate ester of phenytoin that is rapidly cleaved in red blood cells and other organ systems to phenytoin. In contrast to phenytoin, it is rapidly and completely absorbed following intramuscular administration (reaching peak levels in 3 hours). When given intravenously, it can be infused at a rate three times faster than phenytoin. The potential adverse side effects of hypotension and ventricular arrhythmia seen with phenytoin infusions have not been reported with fosphenytoin. Fosphenytoin is administered in units called phenytoin equivalents (PE), which is the amount of phenytoin to be used rather than fosphenytoin itself. Therefore, the dosing of fosphenytoin in status epilepticus is written as 20mg PE/kg and can be administered intravenously at a rate of 3mg PE/kg/min in children.

Over the past year, there has developed a nationwide shortage of injectable phenobarbital and therefore may not be available in the pharmacies of many MEDDAC and MEDCEN institutions. As a result, I propose the following modified protocol for the treatment of status epilepticus in children:

* Lorazepam (*Ativan*) .1mg/kg/dose

* Repeat Lorazepam dose .1mg/kg/dose in 5 minutes if seizure persists

* Fosphenytoin (*Cerebryx*) 20mg PE/kg to infuse at a rate of 3mg PE/kg/min

* Repeat Fosphenytoin dose 10mg PE/kg in 30 minutes if seizure persists

* Diazepam (*Valium*) continuous infusion 2mg/kg/hr if seizure persists

In summary, the general pediatrician needs to be aware of significant changes in the evaluation and treatment of the child who presents with a first seizure with or without fever, the new generation anti-epileptic drugs currently on the market, and the changes in the treatment of status epilepticus due to the development of new medications and the unavailability of established drugs.

(Editor's Note: Dr. Suhrbier has chosen to take up the practice of pediatric neurology in Lake Bluff, IL. His contributions to military pediatrics have been very much appreciated and we wish him success in his new practice.)

Of RADishes and Rum Cake

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Case Study #1

Peter is a 12 year-old white male that was admitted to the adolescent mental health inpatient unit after two years of increasing aggression with violent rages, culminating in destruction of the family's kitchen and a threat to kill his mother with a butcher's knife. Peter was followed by a Psychiatrist in the community, who diagnosed him with "Irritable Depression", and had treated him for over one year

with successive trials of Prozac, Zoloft, Depakote and Risperdal with no improvement. Details from the admission history revealed that Peter was raised from birth in a harsh and neglectful orphanage in Ukraine, and had been adopted at age 10 by his current parents. His parents complained that despite two years of heaping love and attention on Peter, he remained aloof, and refused any physical affection. When the parents approached him affectionately, he would fly into rages resulting in injury to them and destruction of their house. They reported "everything is a power struggle with him" and "no matter what we do, he seems to suspect us of trying to trick him or use him." Despite this, Peter would occasionally seek attention from them, but if the encounter did not go according to Peter's desires, he would again become violent. Peter was also given to stealing, lying, and manipulating at home and at school. Peter characterized his parents as "ok", but that he was "never sure if they [could] be trusted." About his adoptive mother, he reported "she makes a good rum cake" but could say nothing else.

Case Study #2

Mary is a 5 year-old white female that was evaluated as an outpatient for extreme aggression. In the waiting room, she was noted to be a cute little girl with curly hair and a beautiful smile, who was aggressively hitting and pushing her 18 month-old sister. Many of the adults in the waiting room commented to her mother on how adorable she was. In the office, she ran to the doctor, hugged him tightly, and said "I love you, doctor, do you love me", then asked "what's your name?" Her mother described how Mary had never shown consistent affection to her parents, but seemed to instantly bond to total strangers. At 3 years-old, Mary was evaluated for "seeming to be manipulative, intentionally cruel, and violent." At that time her mother was told that this was impossible in a 3 year-old. At 4 years-old, Mary's explosive outbursts and frequent anger were diagnosed as Bipolar Disorder, resulting in a hospitalization and a trial of Depakote, which was ineffective. More revealing was the fact that Mary had been very ill as a baby, requiring repeated, prolonged hospitalizations. Her parents were able to interact with Mary very little until she was nearly two years old, and what few interactions they had were very limited. Since then, the parents had tried very hard to "make up for lost time" and establish loving bonds with her. To their dismay, they discovered that Mary was repulsed by their advances, and seemed to be able to "take us or leave us." Mary also did not recognize their authority in the home and would become rageful if exposed to any boundaries, which resulted in destruction of their home and harm to parents and younger siblings.

Diagnosis

After complete evaluations, these two patients were diagnosed with Reactive Attachment Disorder (RAD) of Infancy or Early Childhood, Disinhibited Type. This diagnosis is suggested by the following criteria (DSM-IV):

A. Markedly disturbed and developmentally inappropriate social relatedness in most contexts, beginning before age 5 years, as evidenced by either (1) or (2):

(1) persistent failure to initiate or respond in a developmentally appropriate fashion to most social interactions, as manifest by excessively inhibited, hypervigilant, or highly ambivalent and contradictory responses (e.g., the child may respond to caregivers with a mixture of approach, avoidance, and resistance to comforting, or may exhibit frozen watchfulness).

(2) diffuse attachments as manifest by indiscriminate sociability with marked inability to exhibit appropriate selective attachments (e.g., excessive familiarity with relative strangers or lack of selectivity in choice of attachment figures)

B. The disturbance in Criterion A is not accounted for solely by developmental delay (as in Mental Retardation) and does not meet criteria for a Pervasive Developmental Disorder.

C. Pathogenic care as evidenced by one of the following:

(1) persistent disregard for the child's basic emotional needs for comfort, stimulation, and affection

(2) persistent disregard of the child's basic physical needs

(3) repeated changes of primary caregiver that prevent formation of stable attachments (e.g., frequent changes in foster care)

D. There is a presumption that the care in Criterion C is responsible for the disturbed behavior in Criterion A (e.g., the disturbances in Criterion A began following the pathogenic care in Criterion C).

Specify type:

Inhibited Type if Criterion A1 predominates in the clinical presentation

Disinhibited Type if Criterion A2 predominates in the clinical presentation

Primacy of Bonding

There is no denying that the inherent strength of the early maternal-child bond is the foundation upon which all future attachments are based. In essence, this diagnosis presumes that a disruption of this important bond has occurred,

resulting in a child's conviction that they cannot depend on others to care for them or meet their needs, which, in turn, results in a disturbance in social relatedness that is present in all settings. Instead of developing bonds of trust, they develop a sense that they are alone in their fears and insecurities, and become manipulative, superficial, and aggressive. Because of this, people who try to draw close to these children are strongly, even violently repulsed[1].

Precipitants

Even after this diagnosis was codified in the *Diagnostics and Statistics Manual of Mental Disorders, fourth edition (DSM-IV)*[2], there remained a pervasive belief among physicians that this diagnostic group was composed mainly of children dying of severe neglect, and therefore it has been linked largely to a diagnosis of Failure-to-Thrive. Ongoing studies at the Attachment and Bonding Center of Ohio, and the Attachment Center at Evergreen, in Colorado, suggest that severe disorders of attachment can result from many pathologic situations, including interruptions in the early bonding cycle due to parental absence, multiple foster home placements, frequent separations such as hospitalization for chronic or recurring illness, abuse or neglect, severe trauma or exposure to trauma, painful medical conditions, chaotic family situations, and lack of nurturing from parents (such as in schizophrenic or depressed parents)[3].

Resultants

Common symptoms that result from these disruptions in bonding include: superficial charm, lack of eye contact when required by parent, indiscriminate affection, not affectionate on parent's terms, destructive to self, others, and property, cruel to animals, stealing personal items, lying about the obvious, poor impulse control, lack of cause and effect thinking, seem to lack a conscience, abnormal eating patterns, cannot make/maintain friendships, incessant chatter, inappropriately clingy or demanding, or mumbling[3]. These symptoms reveal the various aspects of the lack of attachment, including poor relatedness, poor impulse control, and various types of interpersonal manipulation.

Detractors

As in the case studies above, these children are frequently given primary diagnoses of Attention-Deficit/Hyperactivity Disorder, Conduct Disorder, Oppositional Defiant Disorder, Depression, Bipolar Disorder, Intermittent Explosive Disorder, and Pervasive Developmental Disorder. The treatments for these disorders, of course, do not address the core attachment problem, and frequently result in multiple medication trials with lack of improvement, a chronic deteriorating course, frustration for doctors and parents, and ultimately in foster placement or repeated admissions to hospice care.

Treatment: Therapy

Multiple therapies have been developed to address issues specific to RADishes (the affectionate term that parents and therapists use to refer to these kids). Common features of these therapies are: high energy, intense focus, close physical proximity, frequent touch, confrontation, movement, nurturing, constant eye contact, and fast-moving verbal exchanges. Therapies that are detached, non-directive, and aloof have proven ineffective. Examples of these therapies include "Holding Therapy", also known as "Rage Reduction Therapy", "Attachment Enhancement", Intensive Family Therapy, Supportive Therapy, and Educational Therapy. These therapies attempt to help the child work thru their attachment rage (re-parent them), educate and provide skills for parents and others involved with the child, then "hand off" the child to the parents through joint sessions[4].

Treatment: Medications

Pharmacotherapy, as an adjunct to other therapies, may also be helpful. Medications are not targeted at the primary diagnosis of RAD, but instead attempt to lessen some of the more intense symptoms associated with RAD, and treat any identified co-morbidities. While the incidence of RAD and its comorbidities is not known, clinical evidence exists that these children may develop depressive disorders or cycling mood disorders, anxiety disorders, obsessive-compulsive disorders (especially surrounding food), as well as attention-deficit disorders[5]. Pharmacotherapy must take into account that medications that may disinhibit children may make the RAD symptoms precipitously worse. Therefore, benzodiazepines must be avoided. It has also been my experience that RAD children tend to have more side effects and paradoxical reactions to medications than their peers, so the old adage "start low and go slow" usually applies. While there are no scientific studies to support their use, the most commonly employed medications for these children are the antipsychotics (e.g., Risperdal) and antiepileptics (e.g., Depakote). These are commonly chosen for their supposed effect in calming aggression, however efficacy and safety data for this use are lacking. In treating comorbid anxiety disorders and depressive disorders, clinical experience suggests avoiding medications that may be activating (e.g., Prozac) in favor of those that tend to be mood-neutral (Effexor).

Use of stimulants for ADHD symptoms should be carefully weighed against the potential for appetite suppression, which may worsen controlling behaviors centered on food, and paradoxical reactions which may worsen their aggression.

Conclusion

As the trend toward adopting children from countries such as Uganda and Ukraine increases, and as psychological studies continue to characterize important clinical issues related to attachment and bonding, it will become increasingly necessary for the primary care doctor to understand these disorders, and be able to actively participate with therapists and parents in their treatment.

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February is American Heart Month and National Children's Dental Health Month

Beyond the Harrington Rod: Modern Philosophy of Scoliosis Evaluation and Treatment for the Non-Surgeon

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Think back to when you learned most of what you "know" about scoliosis. If the answer is more than 10 years ago, then your knowledge is probably significantly out of date. Few aspects of Pediatric Orthopaedics have changed more in the past decade than the approach to the child with scoliosis. The purpose of this article is to review current consensus on the evaluation and treatment of scoliosis in children.

Scoliosis is broadly categorized by type: congenital, neuromuscular, or idiopathic, and then further subcategorized within this framework. This classification scheme is enormously useful since it highlights the differences in pathoanatomy, natural history and most importantly, treatment regimen. However, a more detailed understanding of the classification of scoliosis is scarcely necessary for the non-surgeon. What is required is the accurate *recording of an observation*, and the timely, appropriate *referral to a treating surgeon*.

The first step in the process, "recording of an observation" has undergone significant modification over the past thirty years. The old system of screening the entire school-age population of children by the school nurse with a scoliometer, instituted in 1963, has been discarded. Multiple studies have confirmed that this system is not cost-effective, and therefore most school systems have appropriately discontinued the practice. The question is, "Who has replaced the school nurse?"[1,4]

The answer is the Pediatrician (or his staff) at one of three opportunities: school physicals, sports physicals, or routine appointments. If the reader participates in any of the above encounters, it is important for him to know that there is no longer a system of observation in place below him. The responsibility for initiating a work-up for scoliosis no longer rests with the school system, but rather with the primary care provider.

With that in mind, what is necessary to make a valid observation? First, a basic understanding of the spatial nature of the deformity is required. As has been frequently noted, scoliosis is not a two-dimensional deformity. Traditionally, scoliosis has been represented in textbooks by a picture or radiograph of a child from the ventral or dorsal perspective. This has unfortunately led to the misunderstanding of scoliosis as primarily, or even predominantly, a coronal plane deformity. In other words, scoliosis is frequently thought of as an improper *bend* in the spine. In fact, scoliosis is never a two-plane deformity. Scoliosis can much more accurately be thought of as a *twist* in the spine, resulting in both coronal and sagittal plane deformity.

The physical manifestations of this twisting of the spine are present in all patients with scoliosis, but to variable degrees. Accurate measurement of the degree of deformity in multiple planes by either physical exam or radiographs is not necessary prior to the evaluation by an orthopaedist. All that is necessary is a physical exam that measures

pelvic and trunkal level and balance, gross appearance of the spine and scapula in the erect and forward leaning position, and a gross neurologic exam.[4]

Radiographs are not recommended at the primary care level because of the likelihood that they will be done incorrectly and need to be repeated at the Orthopaedist's office, exposing the child to unnecessary radiation. This is a common mistake. The Scoliosis Research Society now recommends that initial radiographs be taken in the *postero-anterior* and *lateral* planes, rather than *antero-posterior* with or without a lateral view. AP radiographs expose the breast buds to additional radiation and project the vertebral bodies at the incorrect magnification. AP radiographs are appropriate only for the nonambulatory child who must sit or lay supine for the radiographic exam. The lateral view gives important information about the sagittal plane deformity. Once the initial radiographs are obtained, patients can be followed with PA views only until an intervention is prescribed.

Most scoliotic curves have structural and nonstructural (or compensatory) components. Unless the structural components end well before the anatomic ends of the spine, most children will exhibit some degree of decompensation of the spine. The pelvis and/or shoulders may be slightly tilted. The shoulders may be shifted toward the right or left so that they are not centered over the intergluteal cleft. The patient may also seem to have a "flat back" due to the loss of normal thoracic kyphosis and lumbar lordosis.

By placing the patient in the forward leaning position and observing the spinous processes and paraspinal musculature, the examiner can easily see the rotation inherent in a twist of the spine. A device frequently used to quantify the magnitude of rotation, called a scoliometer, can be used to determine this. Rotation of seven degrees has traditionally been considered sufficient to warrant orthopaedic consultation. This number should not be relied on exclusively because some curves with equal coronal plane deformity may have different degrees of rotation. It is therefore important to consider this element of the exam in relation to other observations.

Examination of the neurologic system at the primary care level is straightforward. Standard motor, sensory, and reflex testing for primarily the lower extremities presents little challenge. The key is to also observe the patient during gait. Priority for gait in the human neurologic system is for maximal efficiency of locomotion. Careful coordination between multiple motor groups that cross multiple joints requires significant cerebral and cerebellar interaction. Any deficiency in the functioning of the entire system shows up readily in the gait pattern. Fortunately, it is very easy for even the layperson to observe gait deficiencies. While the accurate characterization for the etiology of the gait abnormality may be difficult, it is also unnecessary at the primary care level. Observation of a nonspecific gait abnormality combined with other physical exam findings suggestive of scoliosis should be enough to warrant the suspicion of neuromuscular scoliosis and the generation of a consult to the Pediatric Orthopaedist.

The treatment of scoliosis has made truly spectacular advances over the past 35 years. Operative treatment of scoliosis began with fusion without instrumentation about one hundred years ago. In fact, the symbol of Orthopaedic Surgery, the sapling tied to the staff, is meant to represent treatment of scoliosis. "Orthopedia" in Latin means, of course, "straighten the child". Both the discipline's name and the use of the symbol are attributed to Nicholas Pare, a French physician, in 1741.

The greatest problem with fusion without instrumentation is that it required immobilizing the patient in a body cast and inpatient admission to the hospital for up to one year. One of the great advances in medicine was the introduction of the Harrington rod in the 1960's. The Harrington rod was fastened to the posterior elements of the spine by hooks and then used to generate distraction of the concave portion of the spine, thus lengthening and straightening the spine while fusion was occurring. Unfortunately, Harrington rods failed to recognize the spatial nature of the deformity and thus created a new deformity in the sagittal plane which came to be recognized as "flat back syndrome." Harrington rod and hook instrumentation was supplanted in favor of sublaminar wire fixation, pioneered by Luque of Mexico City in the 1970's. Luque fastened precontoured knurled rods to the posterior elements of the spine with sublaminar wires at each level, thus pulling the spine into the three-dimensional shape that the surgeon desired.[3]

A troubling complication, unacceptably high incidence of neurologic injury, combined with the development of a new system by Cotrel and Dubousset in France in the 1980's introduced the next generation of instrumentation method. The CD method of spine deformity correction reverted to hooks and precontoured rods which were used to convert coronal plane deformity into sagittal plane kyphosis and lordosis by *derotating* the spine. The ultimate failure of this system alone was its inability to

place the thoracic kyphosis and lumbar lordosis at the anatomically correct levels. The great contribution of CD technique was the introduction of the concept of *segmental fixation* which allowed infinite combinations of distraction, compression, and derotation at many points along the same rod.[5,6]

The current favored techniques for posterior spinal fusion and instrumentation use hybridized constructs that employ parts of each of the above-mentioned methods. Wires, hooks, and pedicle screws are used to rigidly fix rods in multiple locations to the spine, resulting in highly efficient bi-planar translation and derotation of the spine to near anatomic position, resulting in restoration of both the normal coronal and sagittal contour of the spine.[2]

Experimental methods being explored include thoracoscopic release of disks, anterior placement of cages to maintain anterior column length, gradual correction of scoliosis through the use of staples to cause hemiepiphyseodesis, and titanium expandable ribs. The operative treatment of scoliosis is indeed in a time of unparalled progress that is sure to result in better correction of deformity through progressively lower morbidity procedures.

The evaluation and treatment of scoliosis in children has undergone significant change over the past 10-15 years and continues to progress at incredible speed. The pediatrician now shoulders a larger burden for the identification of patients with possible scoliosis, but the Pediatric Orthopaedist stands ready to complete the evaluation and initiate treatment when necessary. Continued research has led to treatment algorithms which are much better tolerated by the child than in the past and can be expected to yield superior results. Considerable optimism exists for newer, less invasive techniques that are being tested at multiple centers now.

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March 17-23 is National Poison Prevention Week and National Inhalants and Poisons Awareness Week

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